

# The Laws . . .

A person transporting a child shall secure the child in a safety seat according to the vehicle and the child safety seat manufacturers' instructions if the child is under the age of **four years** (regardless of the child's weight), or if the child weighs **forty pounds** or less, regardless of the child's age.

## Maryland's Child Passenger Safety Laws

**\*This means that a child must be properly restrained in a federally approved child safety seat until he/she has achieved both four years of age AND weighs forty pounds.**

A person may not transport a child younger than **sixteen years** unless the child is secured in a child safety seat or a vehicle seat belt.

A child younger than **sixteen years** may not ride in the unenclosed cargo bed of a (Class E) pickup truck

### Important Tips to Remember



- It is the driver's responsibility to ensure that each child is properly restrained.
- Failure to obey the child passenger safety law is a primary offense. You can be pulled over and fined \$40 if any child is not properly buckled.
- Passenger cars, multi-purpose vehicles, and light duty trucks are covered by the law.
- Safety seats and seat belts must be used correctly. Buckle only one child in each seat belt.
- Booster seats are considered safety seats. For your child's safety, KISS recommends keeping children in boosters until they reach 60 pounds.

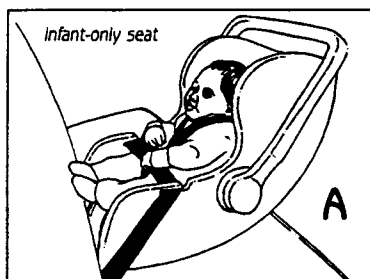
**If you cannot afford a safety seat,  
call Maryland K.I.S.S. at 1-800-370-SEAT.**

### Maryland Adult Seat Belt Law

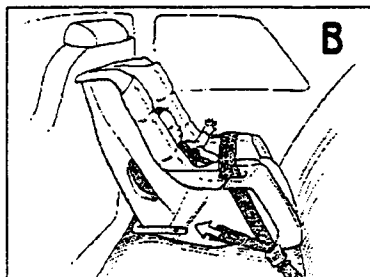
Drivers must wear seat belts and ensure that front seat passengers under **sixteen years of age** wear seat belts or be secured in safety seats.

Front seat passengers age **sixteen years** or older must wear seat belts.

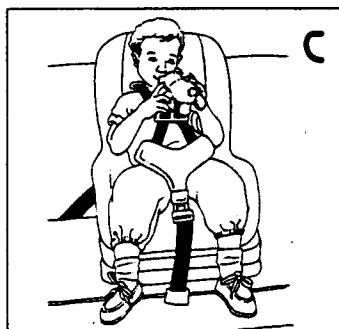
Failure to obey this law is a primary offense. You can be pulled over and fined \$25.



Infants under 1 year and less than 20 lbs. face rear only.



Infants less than 1 year, over 20 lbs. ride in a seat approved for heavier infants rear facing.



Child over age 1 and at least 20 lbs faces the front.



Belt-positioning booster is used with both lap and shoulder belts.

Auto booster seats are for children who have outgrown safety seats, at about 40 pounds.



# Tip #1

## quick safety seat checkup

### Does your child ride in the back seat?

- The back seat is generally the safest place in a crash.
- If your vehicle has a passenger air bag, it is essential for children 12 and under to ride in back.

### Does your child ride facing the right way?

- Infants should ride in rear facing restraints, preferably in the back seat, until about age 1 and at least 20-22 lbs (A). Infants who weigh 20 lbs. before 1 year of age should ride in a restraint approved for higher rear facing weights (B). Always read your child restraint owner manual for instructions on properly using the restraint.
- Children over age one and at least 20 pounds may ride facing forward (C).

### Does the safety belt hold the seat tightly in place?

- Put the belt through the right slot. If your safety seat can be used facing either way, use the correct belt slots for each direction.
- The safety belt must stay tight when securing the safety seat. Check the vehicle owner's manual for tips on using the safety belts.

### Is the harness buckled snugly around your child?

- Keep the straps over your child's shoulder. The harness should be adjusted so you can slip only one finger underneath the straps at your child's chest. Place the chest clip at armpit level.

### Does your child over 40 pounds have the best protection possible?

- Keep your child in a safety seat with a full harness as long as possible, at least until 40 pounds (C). Then use a belt-positioning booster seat which helps the adult lap and shoulder belt fit better.
- A belt-positioning booster seat is preferred for children between 40-80 pounds (D). It is used with the adult lap and shoulder belt. Check on special products for heavy children too active to sit still in a booster.

### How should a safety belt fit an older child?

- The child must be tall enough to sit without slouching, with knees bent at the edge of the seat, with feet on the floor. The lap belt must fit low and tight across the upper thighs. The shoulder belt should rest over the shoulder and across the chest (E). Never put the shoulder belt under the arm or behind the child's back. The adult lap and shoulder belt system alone will not fit most children until they are at least 4'9" tall and weigh about 80 pounds.

For more information, read Child Auto Safety Tips #2 to #9 and call your local safety group or the DOT Auto Safety Hotline: 1-888-DASH-2-DOT.

Even the "safest" seat may not protect your child if it isn't used correctly.



MARYLAND STATE HIGHWAY ADMINISTRATION, MARYLAND COMMITTEE FOR SAFETY BELT USE,  
MARYLAND SAFE KIDS COALITION, AMERICAN ACADEMY OF PEDIATRICS – MARYLAND CHAPTER,  
EMERGENCY NURSES CARE, AMERICAN TRAUMA SOCIETY – MARYLAND DIVISION,  
MARYLAND INSTITUTE FOR EMERGENCY MEDICAL SERVICES SYSTEMS, MARYLAND KIDS IN SAFETY SEATS

## *Prescription for Your Child's Safety From Your Health Care Provider*

PATIENT'S NAME: \_\_\_\_\_ AGE \_\_\_\_\_ WEIGHT \_\_\_\_\_ HEIGHT \_\_\_\_\_

PARENT'S NAME: \_\_\_\_\_

### **R** FOR SAFE TRAVEL IN A MOTOR VEHICLE:

- ☐ *Is your child younger than one year and weighs less than 20-22 pounds?*  
Restrain him or her in an infant or convertible safety seat, rear-facing; NEVER in the front seat with an airbag.
- ☐ *Is your child younger than one year and weighs more than 20-22 pounds?*  
Restrain him or her in a convertible safety seat rated for higher weights rear-facing; NEVER in the front seat with an airbag.
- ☐ *Is your child older than one year and weighs between 20-22 and 40 pounds?*  
Restrain him or her in a convertible safety seat, forward-facing; for older children in the 30-40 pound range, a high-backed booster WITH harness is acceptable.
- ☐ *Is your child less than 10 years of age? Does your child weigh between 40 and 80 pounds, and his/her knees do not bend over the edge of the vehicle seat?*  
Restrain him or her in a booster seat (high-backed for vehicles with low bench seats, no-backed for seats with headrests) used with both lap and shoulder belts.
- ☐ *Does your child weigh more than 80 pounds?*  
Restrain him or her in a lap and shoulder belt.
- ☐ **Do YOU buckle up properly on every ride?**

REFILL: BUCKLE UP ON EVERY RIDE!!

MAKE SURE YOUR CHILD'S SAFETY SEAT IS INSTALLED ACCORDING TO THE MANUFACTURER'S INSTRUCTIONS!  
**KEEP CHILDREN IN THE BACK SEAT UNTIL THEY ARE 13 YEARS OF AGE AND WEIGH AT LEAST 100 POUNDS!**

*For more information, please call Maryland Kids In Safety Seats (KISS) at 1-800-370-SEAT, the  
Maryland State Highway Administration at 410-787-4077, or the Maryland Committee for Safety Belt Use at 410-787-5893.*

# FOOD GUIDE PYRAMID

*A Guide to Daily Food Choices*

COMPLIMENTS OF:

**Fats & Sweets  
USE SPARINGLY**

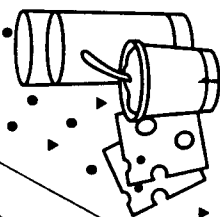
## KEY

These symbols show fats and added sugars in foods.

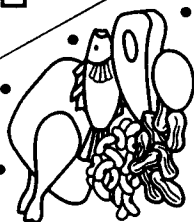
- Fat (naturally occurring and added)
- ▼ Sugars (added)

The Pyramid is an outline of what to eat each day. It's not a rigid prescription, but a general guide that lets you choose a healthful diet that's right for you. The Pyramid calls for eating a variety of foods to get the nutrients you need and at the same time the right amount of calories to maintain a healthy weight.

**Milk, Yogurt, & Cheese Group  
2-3 SERVINGS**



**Meat, Poultry, Fish, Dry Beans, Eggs, & Nuts Group  
2-3 SERVINGS**



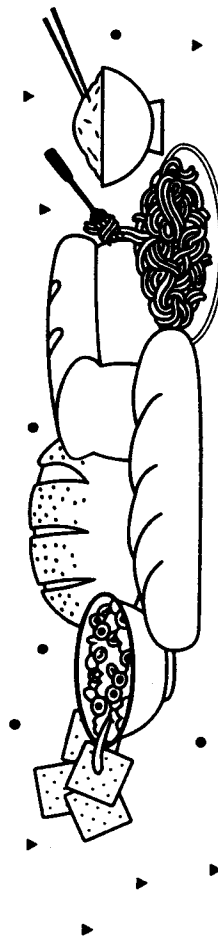
**Vegetable Group  
3-5 SERVINGS**



**Fruit Group  
2-4 SERVINGS**



**Bread, Cereal, Rice, & Pasta Group  
6-11 SERVINGS**



The Food Guide Pyramid emphasizes foods from the five food groups shown in the three lower sections of the Pyramid.

Each of these food groups provides some, but not all, of the nutrients you need. Foods in one group can't replace those in another. No one food group is more important than another—for good health, you need them all.

Source: U.S. DEPARTMENT OF AGRICULTURE and the U.S. DEPARTMENT OF HEALTH AND HUMAN SERVICES.

## What Counts as a Serving?

Serving sizes from each food group vary depending upon the age of the child. These servings sizes may be used as a guide.

Breads, Cereals, Rice & Pasta Group (6 to 11 servings)		1 to 3 years	4 to 6 years	7 and older
Bread, whole grain or enriched		½ to 1 slice	1 slice	1 slice
Tortilla, small muffin, biscuit		½ to 1	1	1
Cooked cereals, rice, pasta		1/4 to 1/3 cup	½ cup	½ cup
Ready to eat cereal		½ cup	½ to 3/4 cup	1 ounce
Vegetable Group (3 to 5 servings)				
Cooked (or cut up raw)		1/4 cup*	½ cup	½ cup
Juice		1/4 cup	½ cup	3/4 cup
Leafy, raw		½ cup *	½ to 1 cup	1 cup
Fruit Group (2 to 4 servings)				
Cooked or canned		1/4 cup	½ cup	½ cup
Whole fruit		1/4 to ½	½	1
Juice		1/4 cup	½ cup	3/4 cup
Milk, Yogurt, & Cheese Group (2 to 3 servings)				
Milk or Yogurt		½ cup	½ to 1 cup	1 cup
Cheese		3/4 ounce	1 ½ to 2 ounces	1 ½ to 2 ounces
Meat, Poultry, Fish, Dry Beans, & Eggs Group (2 to 3 servings) For children ages 7 and older, consume a total of 5 to 7 ounces.				
Meat, poultry, fish cooked		1 ounce	1 to 2 ounces	2 to 3 ounces
Egg		1	1	1 (= 1 ounce)
Peanut butter		*	2 tablespoons	2 tablespoons (= 1 ounce)
Dry Beans, cooked		1/4 to ½ cup	½ cup	½ cup (= 1 ounce)

\* Raw vegetables and peanut butter can be a choking hazard for children ages 2 and under. Wait until the child is older to offer these foods.

## ***BREASTFEEDING PROMOTION***

Breastfed infants should be seen within one week after birth to evaluate both mother and infant for breastfeeding adequacy, to provide anticipatory guidance, and to address practices for maximizing breastfeeding success. Encourage the mother to breastfeed her infant on demand, yet provide a minimum of 8 feedings in 24 hours. Adequate intake can be measured by weight gain, number of wet diapers, and stool output. An infant (especially a newborn) may adequately nurse from a single breast at each feeding. Mothers should offer alternate breasts at subsequent feedings. An older infant often prefers to take both breasts at each feeding. In either case, mothers should feed from both breasts equally during the course of the day.

Changes in frequency of breastfeeding usually reflect a growth spurt and are normal. Counsel mothers to expect this and that by feeding on demand, the milk supply will increase to support the growth of her infant. After 4 months of age, an exclusively breastfed infant's weight percentile on the NCHS growth chart may shift down somewhat from the original growth percentile. This pattern appears to be normal for breastfed infants and does not necessarily indicate insufficient breast milk or inadequate growth progress. Evaluate infants who do not appear to be thriving on breast milk alone or if the mother verbalizes concerns or problems. Use the ***Breastfeeding Observation and Assessment Form*** for completing an evaluation. Provide breastfeeding mothers with the name and phone number of a lactation consultant in the community who can provide additional support.

### **Nutrition Resources and Referral information:**

- Children up to 5 years of age may be eligible for WIC, the Special Supplemental Nutrition Program for Women, Infants, and Children. This federal program provides nutritious foods and nutrition education, including breastfeeding counseling and support. Use the WIC Program Referral Form (in the Patient Care Forms section) or call 1-800-242-4WIC (inside Maryland only) or 1-410-767-5233 (outside of Maryland) to refer patients. Physician Referral Forms and other materials may be ordered. See the WIC Order Form (in the Patient Care Forms section).
- Lactation consultants can provide counseling and support to breastfeeding mothers. They can be found by contacting local hospitals, local health departments, and the Maryland WIC Program.

**MARYLAND  
FAMILY PLANNING  
CLINICAL GUIDELINES**

**1997**

**OFFICE OF MATERNAL HEALTH AND FAMILY PLANNING  
DEPARTMENT OF HEALTH AND MENTAL HYGIENE**

## EMERGENCY CONTRACEPTION

### Rationale

Emergency contraception is contraception used after sexual intercourse but before a woman becomes pregnant. It may be appropriate for women to use emergency contraception when condoms break, diaphragms or cervical caps become dislodged, birth control pills are lost or forgotten, IUD's are expelled, teratogens are taken, no contraceptive is used, or sexual assault occurs. It is an important contraceptive option for women who have had unprotected intercourse within the last 72 hours. Nearly 2 million unplanned pregnancies could be prevented each year and the number of abortions could be cut by half if emergency contraception were widely used.

The main effect of emergency contraception is to prevent pregnancy. It does not cause an abortion. If the initial dose is given within 72 hours of unprotected intercourse, the pills temporarily disrupt ovarian hormone production which may delay or inhibit ovulation. This disruption of ovarian hormone production may cause an inadequate or absent luteal phase which in turn does not allow the endometrium to develop well enough to support implantation. Also, hormone disruption may cause disordered tubal transport.

The pregnancy rate after emergency contraception is less than 2%. If treatment is initiated more than 72 hours after unprotected intercourse, it may not be effective at all.

Previously, combination oral contraceptive pills (eg. Ovral, Triphasil) were the most readily available alternative emergency contraceptive. Research has been done only on norgestrel products so that even though other pills may be effective, they are not currently used. Preven is approved and marketed specifically for emergency contraception.

By using emergency contraception, family planning can be made available to a large group of individuals who are ordinarily outside the system. It is the only way to reduce pregnancy risk in circumstances of rape, mechanical failure, or a lapse in contraceptive protection.

Family planning clinics in the federally funded Title X program received explicit authorization to provide emergency contraceptive treatment in April 1997.

### Plan of Action

1. All women of reproductive age who have had unprotected intercourse in the last 72 hours are eligible. If there has been any other episode of unprotected intercourse since the LMP, emergency contraceptive pills may not be effective since the patient may already be pregnant. Reviewing the cycle time at which exposure occurred may enable the clinician to estimate whether pregnancy risk is high or low, but what determines whether emergency contraception is warranted is how the woman feels about reducing the risk, no matter whether the risk is high or low.



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2. Explore the patient's feelings about continuing pregnancy in the event that emergency contraception does not prevent pregnancy. There is no evidence that emergency hormone contraception would be harmful to a fetus but an attempt should be made to avoid any unnecessary medication during pregnancy.
3. Do a focused evaluation using the Family Planning Emergency Contraception Record-DHMH 4507 (Appendix A). This form should be used for both new and established clients.
4. The urine pregnancy test must be negative.
5. Have the patient sign the informed consent (Consent for Emergency Contraceptive Pills-DHMH 4508) (Appendix B). Give the patient a copy of the instruction sheet (Instruction for Using Emergency Contraceptive Pills-DHMH 4509) (Appendix C) to take home. Place the Family Planning Emergency Record, the signed informed consent, and a copy of the instruction sheet in the chart.
6. If there is no clinician available when a client seeks emergency contraception, the clinic nurse should complete the Family Planning Contraception Record-DHMH 4507 and call a clinician for a telephone order. The order may be faxed for a signature or signed by the clinician at a later date, according to each clinic's administrative guidelines.
7. Five common emergency contraception options:
  - a. Preven - take 2 tablets now and 2 tablets 12 hours later
  - b. Ovral - take 2 tablets now and 2 tablets 12 hours later
  - c. Triphasil - take 4 yellow tablets now and 4 yellow tablets 12 hours later
  - d. Nordette - take 4 tablets now and 4 tablets 12 hours later
  - e. Lo/Ovral - take 4 tablets now and 4 tablets 12 hours later

To avoid making the client take the second dose in the middle of the night, allow her to delay the first dose by a few hours, provided the 72-hour time limit is not exceeded.
8. About 50% of women have nausea and 20% may vomit, therefore, the tablets should be taken with food. Provide anti-nausea medication or recommend an over-the-counter product to be taken 30-60 minutes before starting the emergency contraceptive tablets (Appendix D). Other side effects include headache, breast tenderness, dizziness, abdominal pain, and menstrual cycle disturbances.
9. Remind the client emergency contraceptive pills will not protect her from pregnancy if she has unprotected intercourse in the days or weeks following the treatment. Contraceptive counseling should be done and a method may be initiated according to the guidelines (Appendix E).
10. Tell the patient to expect her menses within 3 weeks.
11. Schedule the patient for a follow-up in 3-4 weeks.

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**Follow-Up**

1. Repeat a pregnancy test at the follow-up visit.
2. If she is not pregnant, counsel the patient and review her contraceptive options.
3. If she is pregnant, inform the patient that although no long term studies have been done specifically to evaluate the risk to the fetus, there is no evidence of any increase in congenital defects among women who take birth control pills and become pregnant. Remind the patient that there is about a 2% incidence of congenital anomalies in all pregnancies.

**Primary References**

ACOG. Emergency Oral Contraception. ACOG Practice Patterns. 1996

Food and Drug Administration. Prescription drug products; certain combined oral contraceptives for use as postcoital emergency contraception. Federal Register, 62:8610-8612, 1997

ACOG. Hormonal Contraception. Technical Bulletin 198. 1994

Hatcher R, et al. Contraceptive Technology. 17th revised edition, Ardent Media, Inc., New York, 1998

Hatcher R, et al. Emergency Contraception: The Nation's Best Kept Secret. Bridging the Gap Communications, Inc., Decatur, GA 1995

Kring T (memorandum). OPA Program Instruction Series, OPA 97-2: Emergency Contraception. 23 April 1997

## **CONSENT FOR EMERGENCY CONTRACEPTIVE PILLS (ECP'S)**

**I request emergency contraceptive pills (ECP's) to minimize a possible pregnancy risk. I understand it is not a main method of birth control.**

**I have received an instruction sheet on how to properly take ECP's.**

**I understand that taking ECP's does not prevent pregnancy 100% of the time. Some pregnancies do occur. In spite of this, I wish to try to prevent pregnancy at this time.**

**I understand that the risk of development of birth defects in the fetus is unknown and that if treatment fails, I must accept this risk should I decide to continue with this pregnancy. No known increased fetal risk of congenital anomalies has been detected so far.**

**I understand that I may not be able to take ECP's if I have had or currently have:**

<b>blood clots in lungs, legs, or eyes</b>	<b>unexplained vaginal bleeding</b>
<b>inflammation of the veins</b>	<b>an already established pregnancy</b>
<b>serious liver disease</b>	<b>cancer of the breast</b>
<b>heart attack or stroke</b>	<b>cancer of the uterus, cervix, or vagina</b>

**I understand that side effects of these pills may include:**

<b>nausea and vomiting</b>	<b>irregular vaginal bleeding</b>
<b>breast tenderness</b>	<b>abdominal pain</b>
<b>headaches and dizziness</b>	<b>menstrual cycle disturbances</b>

**I know to watch for "ACHES" as danger signals and to contact a health care provider immediately if any of these signs occur:**

- A. abdominal pain**
- C. chest pain or shortness of breath**
- H. headache (severe), numbness, or dizziness**
- E. eye problems such as blurred vision or double vision**
- S. severe leg pain**

**I understand that if I see a clinician for any reason before I get my period, I should tell him/her that I have taken ECP's.**

**I understand that I should expect my period within 3 weeks and I agree to have a pregnancy test if it has not occurred within that time. I will also inform a clinician of any lower abdominal pain or bleeding that is unusually heavy or light.**

**Date \_\_\_\_\_ Client Signature \_\_\_\_\_**

**I have reviewed the above information with the client.**

**Date \_\_\_\_\_ Staff Signature \_\_\_\_\_**

APPENDIX D

ANTI-NAUSEA TREATMENT OPTIONS

DRUG	DOSE	TIMING OF ADMINISTRATION
<b>NON-PRESCRIPTION DRUGS</b>		
Meclizine hydrochloride (Dramamine II, Bonine)	One or two 25 mg tablets	1 hour before first ECP dose; repeat if needed in 24 hours
Diphenhydramine hydrochloride (Benadryl)	One or two 25 mg tablets	1 hour before first ECP dose; repeat if needed every 4-6 hours
Dimenhydrinate (Dramamine)	One to two 50 mg tablets or 4-8 teaspoons liquid	30 minutes to 1 hour before first ECP dose; repeat as needed every 4-6 hours
Cyclizine hydrochloride (Marezine)	One 50 mg tablet	30 minutes before ECP dose; repeat as needed every 4-6 hours
<b>PRESCRIPTION DRUGS</b>		
Meclizine hydrochloride (Antivert)	One or two 25 mg tablets	1 hour before first ECP dose; repeat if needed in 24 hours
Trimethobenzamide hydrochloride (Tigan)	One 250 mg tablet or 200 mg suppository	1 hour before first ECP dose; repeat as needed every 4-6 hours
Promethazine hydrochloride (Phenergan)	One 25 mg tablet or suppository	30 minutes to 1 hour before first ECP dose; repeat as needed every 8-12 hours

Source: HATCHER et al. 1998

## **INSTRUCTION FOR USING EMERGENCY CONTRACEPTIVE PILLS**

- 1. Swallow the first dose (e.g. Preven - 2 tablets, Triphasil - 4 yellow tablets, Nordette 4 tablets, Lo/Ovral 4 tablets, or Ovral 2 tablets) as soon as possible and not later than 72 hours after having unprotected sex. Take pills with food so you don't feel too nauseated.**
- 2. Swallow the second dose (e.g. Preven - 2 tablets, Triphasil - 4 yellow tablets, Nordette 4 tablets, Lo/Ovral 4 tablets, or Ovral 2 tablets) 12 hours after taking the 1st dose.**
- 3. You may have been advised to take anti-nausea medication 30-60 minutes before taking the emergency contraception pills. If you feel sick to your stomach, this should only last a day or so. If you vomit in less than 1 hour after taking a dose, inform your clinician. You may need to take additional pills and some anti-nausea medication.**
- 4. Do not take extra pills. More pills are not more effective. They may make you more nauseated and possibly cause you to vomit.**
- 5. Your next period will usually start within 3 weeks. If your period does not start, you could be pregnant and need to have a pregnancy test done. If you have abdominal pain or an unusually light or heavy period, call your clinician for further evaluation. It may be a sign of a more serious condition such as ectopic pregnancy (pregnancy outside the uterus).**
- 6. Side effects of nausea and vomiting are relatively common. Less common side effects include headaches, dizziness, breast tenderness, abdominal pain, and menstrual cycle disturbances.**
- 7. In the next 2 weeks, watch for and report any of the "ACHES" warning symptoms which, though rare, may accompany ordinary birth control pill use:**
  - A. abdominal pain**
  - C. chest pain or shortness of breath**
  - H. headache (severe), numbness, or dizziness**
  - E. eye problems such as blurred vision or double vision**
  - S. severe leg pain**
- 8. Make an appointment to come back for a follow-up visit in 3-4 weeks.**
- 9. From now on, make sure you use birth control every time you have sex. Emergency contraceptive pills are for one-time, emergency use only. They are not as effective nor as easily tolerated as other forms of birth control.**

## APPENDIX E

### INITIATING ONGOING CONTRACEPTION AFTER ECP

Because ECPs can delay ovulation, a client could be at risk of pregnancy in the first few days after treatment. A client should use a back-up method of contraception for the remainder of the treatment cycle then initiate a regular method of contraception with her next menstrual period.

METHOD	WHEN TO INITIATE
Condom	Immediately
Diaphragm	Immediately
Spermicide	Immediately
Oral Contraceptive (OC)	Initiate a new pack, either according to manufacturer's instructions after beginning the next menstrual cycle, or begin taking one OC tablet daily the day after the ECP treatment is completed.  Women using Lo/Ovral, Nordette for emergency contraception can continue taking one pill per day from the same pack.
DMPA	Initiate within 5 days of beginning the next menstrual period.
Norplant	Initiate within 7 days of beginning the next menstrual period.
Intrauterine Device (IUD)	Initiate during the next menstrual period.
Fertility Awareness	Initiate after onset of the next normal menstrual period and after the patient has been trained in using the method.
Sterilization	Perform the operation any time after beginning the next menstrual period.

Adapted from: HATCHER et al. 1998

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**DEPO-PROVERA (DMPA)**

**Rationale**

Depo-Provera, medroxyprogesterone acetate (DMPA), is a long-acting injectable progestin contraceptive which, when given intramuscularly within a 13 week interval, is a highly reliable method of birth control. It inhibits ovulation by suppressing FSH and LH levels, thereby blocking the LH surge and inhibiting ovulation. Other mechanisms of action are thickening of the cervical mucus, creation of a thin atrophic endometrium and premature luteolysis. DMPA is effective immediately after the first injection.

DMPA is administered in a 150 mg dose by a deep intramuscular injection in the deltoid (upper arm) or gluteus (buttock). When given as directed, the first-year probability of failure is 0.3%.

DMPA may well be the ideal contraceptive for women who have difficulty remembering to take pills every day, who dislike using methods associated with coitus, who are not suitable candidate for IUDs or who are not ready to make a five year commitment to Norplant. DMPA is an excellent contraceptive option for women who cannot take estrogen (e.g. congenital heart disease, sickle cell anemia, a previous history of thromboembolism, and women over 30 who smoke). DMPA can be used by breastfeeding women since it does not interfere with lactation once milk flow has been established, nor does it have any observable effects on the infant. Milk flow may actually increase. DMPA should be considered in patients with seizure disorders; an improvement in seizures control can be achieved probably because of the sedative properties of progestins. Other benefits of DMPA use include a decreased risk of endometrial cancer, and probably the same benefits associated with the progestin impact of oral contraceptives: reduced menstrual flow and anemia, less PID, less endometriosis, fewer uterine fibroids, and fewer ectopic pregnancies. There is no increased risk of breast cancer.

Possible side effects of DMPA include irregular bleeding, breast tenderness, weight gain, depression or mood changes, alopecia and decreased libido. Regular clinic visits are required for injections. There is no STD or HIV protection. There is no immediate discontinuation of DMPA and it may take 6-8 months to clear from the body. The return to fertility is about 9 months after the last injection.

**Plan of Action**

1. Clients expressing an interest in this method should have the method, its benefits and its side effects fully explained and be given a patient information brochure to study. The Depo-Provera consent should be signed.
2. Reasons for not using DMPA include:
  - a. Confirmed or suspected pregnancy
  - b. Undiagnosed vaginal bleeding

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- c. Confirmed or suspected breast cancer
  - d. Acute liver disease or tumors
  - e. Acute thrombophlebitis or thromboembolic disease
  - f. Known sensitivity to Depo-Provera
  - g. Current use of aminoglutethamide (used in cancer chemotherapy and Cushing Syndrome)
3. Special conditions for which physician consultation is recommended before starting DMPA:
- a. Severe depression
  - b. Severe migraine headaches
  - c. Severe cardiovascular disease and/or history of heart attack or stroke
  - d. Chronic liver disease
5. DMPA is available in 150 mg vials (1 cc) and in prefilled syringes (available summer 1996). The needle should be 1 1/2" long and 21-23 gauge. The vial should be shaken before administration to insure uniform mixing of the suspension. Deep intramuscular injection may be made into the deltoid or the gluteus maximus muscles. The area of the injection should not be massaged because this may lower the effectiveness of Depo-Provera.
6. New labeling from Upjohn emphasizes the need to determine that the patient is not pregnant before injection, if the interval extends beyond 13 weeks. The order should read: Depo-Provera (or DMPA) 150 mg IM q 12 week or (q 11-13) x 5. Controversy persists concerning how long the contraceptive level is maintained if the interval extends beyond 13 weeks.
7. DMPA may be given earlier than 13 weeks, for circumstances such as the injection being due while the patient is away on vacation or before a college student returns to school. An earlier injection at 11, 10, or even 9 weeks is acceptable; however, the goal is within the 13 week interval.
8. The initial DMPA injection options include:
- a. During the first 5 days of a normal menstrual period and with a negative urine pregnancy test.
  - b. Within 5 days after childbirth for women who are not breastfeeding.



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- c. Anytime after the milk flow has been established for breastfeeding women who have not resumed sexual intercourse.
- d. All other circumstances necessitate that the patient is confirmed not to be pregnant. This may be accomplished by getting a negative urine pregnancy test before and after a 2 week interval of abstinence or use of reliable contraception.
- e. When switching from oral contraception to DMPA, the first injection should be given during the first 5 days of the withdrawal menstrual period or if necessary, due to scheduling problems, anytime during the last pack of pills.
- f. When switching from IUD to DMPA, the first injection may be given during the first 5 days of menstruation and the IUD may be removed immediately; or anytime while the IUD is in place but the IUD must not be removed for 2 weeks after the injection.
- g. The first DMPA may be given anytime prior to the removal of Norplant.
- h. In unusual situations which may require clinical judgement, consideration should be given to a review of the patient's recent sexual activity, current method of contraception compliance, pelvic examination, urine pregnancy test and/or physician consultation.

**Follow-Up**

- 1. A reminder card may be prepared at the time of injection which lists the scheduled date for the next injection. A copy of this can be carried by the patient and a copy put in a tickler file to be mailed to the patient as a reminder 2 weeks before the expected date.
- 2. When a patient is late for her next DMPA injection (more than 13 weeks since the last injection), pregnancy must be ruled out before the next dose is given. This may be accomplished by getting a negative urine pregnancy test before and after a 2 week interval of abstinence or use of reliable contraception.
- 3. Management of side effects:
  - a. Menstrual Changes - Almost all women experience spotting, irregular or prolonged bleeding during the first few months of DMPA use. Amenorrhea occurs in about half of the women by the end of the first year and increasingly thereafter. The patient should be reassured and may be given 1 or 2 cycles of a low-dose combined oral contraceptive to control bleeding. Another option is Ibuprofen 200-400 mg q4h x 5 days. Prolonged or excessive bleeding requires physician consultation.

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- b. **Weight Gain** - The average weight gain can be 4-5 pounds per year over the first 3 years. The weight usually stabilizes.
- c. **Amenorrhea** - Cessation of menses is expected over time in most women on DMPA. However, if the onset is immediate, pregnancy must be ruled out. There is no need to induce menses.
- d. **Bone Loss** - There is some degree of bone loss due to lower estrogen levels. It is unlikely that this bone loss is sufficient to raise the risk of osteoporosis later in life. Bone density measurements in women who stopped using DMPA indicated the loss is regained even after long-term use.
- e. **Other possible side effects** include headaches, breast tenderness, loss of libido, depression, nervousness, and fatigue.

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**PRECONCEPTION COUNSELING**

**Rationale**

Preconception counseling offers women an ideal time to plan their pregnancies and establish good health habits. Certain congenital anomalies and complications of pregnancy may be prevented if intervention occurs prior to conception. Fetal organogenesis begins 17 days after fertilization and is completed by the time many women have their first prenatal appointment. Promoting positive health behaviors and eliminating medical risks are most effective when initiated preconceptionally.

Since up to 60% of all pregnancies are unintended, targeting only self-referred women who are planning their next conception will result in a significant number of missed opportunities for primary prevention. Counseling women of childbearing age allows for an identification of women with risk factors. As an example, we can educate women to avoid any teratogenic medications, get immunized to rubella, and take folic acid supplements to decrease their risk of neural tube defects. The active planning of pregnancy will maximize the benefits of appropriate interventions and adherence to good health habits to help insure a reduction of maternal and perinatal morbidity and mortality.

**Plan of Action**

1. A systematic identification of preconception risks should be offered through assessment of reproductive, family and medical histories, nutritional status, drug exposures, and social concerns of all fertile women (Appendix A). The preconception questionnaire (Appendix B) or the March of Dimes questionnaire may be utilized.
2. Educate and counsel the patients based on the identified risks.
3. Discuss the possible effects of pregnancy on existing medical conditions and offer medical consultation.
4. Discuss genetic concerns and refer if appropriate.
5. Determine immunity to rubella, varicella, and hepatitis. Immunize if indicated.
6. Recommended laboratory tests are essentially the same as those obtained at the first prenatal visit.
  - a. hgb/hct
  - b. blood type and Rh
  - c. antibody screen
  - d. rubella titer
  - e. hepatitis screen (HBsAg)
  - f. Pap, GC, CT, STS
  - g. PKU
  - h. urinalysis

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- i. hemoglobin electrophoresis
  - j. glucose screening, if indicated
  - k. PPD, if indicated
  - l. HIV, counsel and recommend screening
7. Nutritional counseling should include a review of appropriate weight for height, sources of folic acid, avoidance of vitamin oversupplementation, and eating a well-balanced diet. Refer for in-depth counseling if appropriate. The U.S. Public Health Service recommends daily supplementation with 0.4 mg of folic acid for all U.S. women of childbearing age to reduce the risk of neural tube defects. Virtually all multivitamins contain 0.4 mg of folic acid. Also many breakfast cereals contain large amounts of folic acid - a bowl of Product 19 or Total satisfies the daily folic acid requirements. Fortunately, the FDA will begin fortification of all grains in 1998, but until then, taking a vitamin or eating food rich in folic acid such as some of the breakfast cereals, may be the easiest way to insure women get their daily quotient. Women who have had a previous pregnancy complicated by a neural tube defect should consume 4.0 mg a day of folic acid when they are planning to conceive. This dose is available by prescription only. Caution must be used when prescribing folic acid because it makes the diagnosis of a vitamin B-12 deficiency (with its neurologic sequelae) more difficult in certain individuals, such as the elderly.
8. Discuss social, financial, and psychologic issues in preparation for pregnancy.
9. Discuss contraceptive use and birth spacing. Encourage the use of a menstrual calendar.
10. Emphasize early and continuous prenatal care.
11. Encourage patients to minimize or avoid their use of caffeine, alcohol, cigarettes, street drugs, and to take appropriate precaution against occupational hazards. Advise patients on available resources (Appendix C).
12. Discuss handling of cats and litterboxes, and dangers of eating undercooked meat to avoid toxoplasmosis.
13. Patients identified with social, psychological, medical and/or genetic risks should be referred appropriately.
14. Counsel patients to avoid hyperthermia in the 1st trimester. Since a maternal core temperature over 100°F. has been associated with birth defects, patients should limit sauna and hot tub sessions to 15 minutes and also limit strenuous exercise (eg. marathon running) in the 1st trimester.
15. Review all medications, including prescribed, vitamins, and over-the-counter. Review the workplace and household for potential hazards to pregnancy. Discuss teratogenicity and toxicity of harmful agents.

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**APPENDIX A**

**PRECONCEPTION RISK FACTORS**

- 1. Medical**
  - a. Chronic conditions such as diabetes, epilepsy, hypertension, thyroid disease, anemia, hepatitis, pelvic infections, lupus, deep vein thrombosis, PKU, asthma, cancer, AIDS, kidney disease**
  - b. Medications - over-the-counter, including vitamins; prescription**
  - c. Lack of immunity to rubella, hepatitis**
  - d. Ethnic origin of Black, Mediterranean, SE Asian, Ashkenazic Jewish**
  - e. Family history of birth defects, mental retardation, or genetic abnormalities**
- 2. Obstetric History**
  - a. Previous stillbirth, neonatal death, miscarriages, need for neonatal intensive care**
  - b. Previous child with birth defect, mental retardation or genetic abnormality**
  - c. Delivery of a child >9lbs. or <5.5lbs.**
  - d. History of diabetes or hypertension during pregnancy**
- 3. Life Style**
  - a. Cigarettes, alcohol, caffeine, street drugs**
  - b. Close association with cats**
  - c. Sauna or hot tub use**
  - d. Poor nutrition**
  - e. Exposure to lead, mercury, radiation, chemicals, or other toxins**
  - f. Occupational hazards**
  - g. Psychosocial stress**
  - h. Domestic violence**

**APPENDIX B**

**PRECONCEPTION HEALTH QUESTIONNAIRE**

<b>LIFESTYLE - PLEASE CHECK ALL THAT APPLY</b>	
1. <i>Indicate how much or how often you:</i>	
a. smoke cigarettes ____	b. drink wine, beer, liquor ____
c. drink coffee, tea, soda ____	d. use marijuana, cocaine, or any other street drugs ____
e. own or work with cats ____	f. use sauna or hot tubs ____
2. <i>Your normal daily diet DOES NOT include:</i>	
2 servings of milk/dairy product ____	2 servings of meat/fish/poultry/eggs/cheese ____
4 servings of fruits/vegetables ____	4 servings of breads/cereals ____
3. <i>In your job or home, do you have contact with:</i>	
a. lead (solder, pipes, batteries, paints, ceramics, emissions) ____	
b. mercury (thermometers, dyes, inks, pesticides, dental fillings) ____	
c. other chemicals (dry cleaning fluids, pesticides, textiles, plastics) ____	
d. radiation ____	
4. Do you feel ready to support a child emotionally and financially? ____ (yes or no)	
5. Has anyone important to you (boyfriend, parent, partner, family member, friend) physically hurt you, and/or forced you to perform sexual acts, and/or made you feel afraid? ____ (yes or no)	
6. <i>Ethnic background:</i>	
a. Mediterranean ____	b. Middle Eastern ____
c. S.E. Asian ____	d. French Canadian ____
e. Black ____	f. Ashkenazic Jewish (E.European) ____
7. Are you and your partner related outside of marriage? ____	
8. Will you be over 34 or under 17 at the time of delivery? ____	

**APPENDIX B**

**PRECONCEPTION HEALTH QUESTIONNAIRE**

<b>MEDICAL - PLEASE CHECK ALL THAT APPLY</b>	
9. <i>Check if you or your partner has:</i>	
a. diabetes ____	b. thyroid disease ____
c. PKU (phenylketonuria) ____	d. asthma ____
e. heart disease ____	f. high blood pressure ____
g. sexually transmitted diseases ____	(gonorrhea, syphilis, herpes, warts, AIDS)
h. lupus ____	i. epilepsy ____
j. sickle cell anemia ____	k. depression or other psychiatric disorders ____
l. other ____	
10. List all medications (over-the-counter, prescription, vitamins) you currently take:	
____	
____	
11. <i>Have you ever been tested or immunized for:</i>	
a. German measles (rubella) ____	b. hepatitis ____
c. tetanus ____	d. AIDS ____
e. varicella ____	
12. Do you, your partner, or any relatives, have birth defects, mental retardation, or genetic abnormalities? ____ If so, please list ____	
<b>PREGNANCY HISTORY - PLEASE CHECK ALL THAT APPLY</b>	
13. <i>Have any of your previous pregnancies resulted in:</i>	
a. stillbirth ____	b. neonatal death ____
c. miscarriage ____	d. need for neonatal intensive care ____
e. diabetes or high blood pressure during pregnancy ____	f. child with birth defects, mental retardation, or genetic abnormality ____
g. birthweight less than 5.5 pounds or greater than 9 pounds ____	

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**DOMESTIC VIOLENCE**

**Rationale**

Domestic violence is violent behavior committed by one partner against the other; usually women are the victims. The violence can be physical, sexual or psychological. Its purpose is to control, dominate or hurt another within an intimate relationship. It is primarily a learned pattern of behavior whose effects, without intervention, become more destructive over time. While domestic violence is directed at a particular victim, it also victimizes children, other family members and the community. Young women may experience this type of abuse in their dating relationships.

Abuse of a woman by an intimate male partner is related to child abuse in several important ways. Half of all men who abuse their female partners also abuse their children and one fourth of the women also abuse the children. Children may come to believe that physical violence is an acceptable way of dealing with problems or conflicts. Also, children who are abused are likely to enter into abusive relationships when they become adults.

Medical professionals need to view physical abuse as a health problem. Appropriate health care management of all women includes universal screening for domestic violence.

Many abused women are reluctant or unable to get help. Some are literally held captive, others may not have the money or means of transportation to leave. Cultural, ethnic and/or religious background may also influence a woman's response to abuse and her awareness of viable options.

Maryland does not have mandatory reporting requirements for domestic violence. It is the responsibility of the victim to report the abuse to the proper authorities. However, whenever any health care provider treats a person for an injury which was caused by or shows evidence of having been caused by any type of gunshot, the police must be notified. In Maryland, mandatory reporting is required for child abuse, abuse of institutionalized individuals, abuse of developmentally disabled individuals, and abuse of a vulnerable adult.

**Plan of Action**

1. All women should be educated to increase their awareness of violence as a health problem. This information should be part of their health care education and counseling.
2. Family planning patients should be screened for the possibility of domestic violence. In addition, information concerning a woman's experience with abuse as a child is important because of the impact on her mental and physical health throughout her lifespan. Direct, specific questions should be phrased so as to make them nonjudgmental and nonthreatening. (Appendix A)

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3. Health care providers should accurately document any problems or complaints of abuse, or physical evidence of violence.
4. The possibility of domestic violence should be considered when a woman's explanation of how an injury occurred does not seem plausible, when there has been a delay in seeking medical care, or when a woman presents with vague complaints.
5. Upon determining that a women is abused, the health care provider needs to assess her level of safety and assist the client in establishing a plan to deal with the abusive situation. Clients should be referred for abuse counseling.
6. All clinic service sites should develop and maintain a list of referral sources available locally to assist the victim of abuse, as well as information about legal and criminal prosecution options.
7. Subsequent clinic visits should include inquires and documentation regarding abuse status.
8. Choice of contraception might be affected by an abusive relationship. For example, injectable progestin (Depo-Provera) might be appropriate for the privacy that this method provides.
9. Patients should be advised that pregnancy does not cure domestic violence. In one study physical abuse continued at the same level or at an increased level in 64% of patients during pregnancy.

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APPENDIX A

Figure 1  
Abuse Assessment Screen

1. Have you ever been emotionally or physically abused by your partner or someone important to you? YES NO

2. WITHIN THE LAST YEAR, have you been hit, slapped, kicked or otherwise physically hurt by someone? YES NO

If YES, by whom? \_\_\_\_\_

Total number of times \_\_\_\_\_

If you are currently pregnant:

3. Since you've been pregnant, were you hit, slapped, kicked or otherwise physically hurt by someone? YES NO

If YES, by whom? \_\_\_\_\_

Total number of times \_\_\_\_\_

Mark the area of injury on the body map.

Score each incident according to the following scale:

- 1 = Threats of abuse including use of a weapon
- 2 = Slapping, pushing; no injuries and/or lasting pain
- 3 = Punching, kicking, bruises, cuts and/or continuing pain
- 4 = Beating up, severe contusions, burns, broken bones
- 5 = Head injury, internal injury, permanent injury
- 6 = Use of weapon; wound from weapon



SCORE

\_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_

(If any of the descriptions for the higher number apply, use the higher number.)

4. WITHIN THE LAST YEAR, has anyone forced you to have sexual activities? YES NO

If YES, whom? \_\_\_\_\_

Total number of times \_\_\_\_\_

5. Are you afraid of your partner or anyone you listed above? YES NO

Developed by the Nursing Research Consortium on Violence and Abuse. Readers are encouraged to reproduce and use this assessment tool.

## CHILD ABUSE AND NEGLECT KNOW YOUR RESPONSIBILITY

Child abuse and neglect is a serious problem which requires the involvement of all private citizens and professionals in the community for the purpose of prevention, identification, and treatment. In Maryland, the child abuse and neglect law requires that anyone who SUSPECTS a child has been or is being mistreated must report the matter to the Department of Social Services. (In case of child abuse, a report may be made to Social Services or the Police.) Any professional who knowingly fails to make a required report of child abuse may be subjected to certain professional sanctions. The professionals identified in Maryland Law include: health practitioners, police officers, educators, and social workers. And, any person who, in good faith, makes a report of abuse or neglect is IMMUNE from civil liability or criminal penalty.

The following information is provided so that you will be familiar with the physical and behavioral indicators of child abuse, neglect or mental injury. Please note that the presence of any of these indicators does not necessarily mean a child is being abused, neglected or is a victim of mental injury. They may, however, lead you to suspect abuse, neglect or mental injury and, therefore, to report it.

### PHYSICAL ABUSE

#### Physical Indicators

Bruises: on any infant; facial bruises; in unusual patterns; clustered in one area of the body; various stages of healing; both eyes "blackened" with no injury to the nose.

Burns: caused by immersion in hot liquid; cigarette burns usually on palms of hands (leaving "crater" shaped burns); caused by hot implement, such as an electric curling iron (leaving burn marks in the shape of the implement); or caused by ropes that indicate confinement.

Wells, cuts, abrasions, fractures, and internal injuries may also indicate abuse. Since these injuries may occur through normal childhood experiences, they should only cause concern when coupled with some other physical or behavioral indicator. You should also be concerned if the injury does not seem likely to have resulted from normal activity, given the child's age and physical development.

#### Behavioral Indicators

Child: overly compliant, shy, or aggressive behavior; avoids parents; inhibited crying; hyperactive; avoids physical contact; low tolerance for frustration; distrustful.

Parent: holds unrealistic expectations for the child's physical or emotional development; "immature"; dependent; aggressive; low sense of self-esteem; sees the child as "bad", "different", or "evil"; low tolerance for frustration; inappropriate coping skills.

## NEGLECT

#### Physical Indicators

Child: extremely dirty and unkempt; clothes inadequate for the weather; serious medical problems left untreated; inadequately supervised; undernourished.

#### Behavioral Indicators

Child: withdrawn; shy; passive; always tired; developmentally slow.

Parent: apathetic; shows little concern or awareness of the child's needs; shows anger when questioned about child's care; impulsive in making decisions; inconsistent disciplinary practice; overwhelming personal needs.

## SEXUAL ABUSE

#### Physical Indicators

Child: difficulty in sitting or walking; repeated symptoms of medical problem with genitals or digestive system; presence of sexually transmitted diseases; pregnancy.

#### Behavioral Indicators

Child: unusual sexual behavior or knowledge; nightmares; poor peer relationships; few social skills, extremely isolated; repeated "runaways."

Parent: extremely overprotective; overly interested in child's social and sexual life; sees child as highly sexualized; jealous.

## MENTAL INJURY

#### Behavioral Indicators

Child: any observable, substantial impairment of a child's mental or psychological ability to function that is a direct result of an act or omission by a parent or caretaker. The child may have severe problems in areas of functioning such as family and/or social relationships, sleeping and eating, academics and overall development, and need specific psychiatric, psychological or social work intervention.

Parent: frequently threatens to harm or kill the child, threatens to harm or kill the child's pet, constantly denigrates the child or subjects the child to extensive emotional or physical isolation or confinement.



## Rare and Expensive Disease List as of July 1, 1998

ICD-9 Code	Disease	Age Group	Guidelines
x all	Symptomatic HIV disease/AIDS (pediatric)	0-20	<p>(A) A child &lt; 18 mos. who is known to be HIV seropositive or born to an HIV-infected mother and:</p> <ul style="list-style-type: none"> <li>* Has positive results on two separate specimens (excluding cord blood) from any of the following HIV detection tests: <ul style="list-style-type: none"> <li>--HIV culture (2 separate cultures)</li> <li>--HIV polymerase chain reaction (PCR)</li> <li>--HIV antigen (p24)</li> </ul> </li> </ul> <p>N.B. Repeated testing in first 6 mos. of life; optimal timing is age 1 month and age 4-6 mos.</p> <p style="text-align: center;">or</p> <ul style="list-style-type: none"> <li>* Meets criteria for Acquired Immunodeficiency Syndrome (AIDS) diagnosis based on the 1987 AIDS surveillance case definition</li> </ul>
V08	Asymptomatic HIV status (pediatric)	0-20	<p>(B) A child &gt; 18 mos. born to an HIV-infected mother or any child infected by blood, blood products, or other known modes of transmission (e.g., sexual contact) who:</p> <ul style="list-style-type: none"> <li>* Is HIV-antibody positive by confirmatory Western blot or immunofluorescence assay (IFA)</li> </ul> <p style="text-align: center;">or</p> <ul style="list-style-type: none"> <li>* Meets any of the criteria in (A) above</li> </ul>
795.71	Infant with inconclusive HIV result	0-12 months	<p>(E) A child who does not meet the criteria above who:</p> <ul style="list-style-type: none"> <li>* Is HIV seropositive by ELISA and confirmatory Western blot or IFA and is 18 mos. or less in age at the time of the test</li> </ul> <p style="text-align: center;">or</p> <ul style="list-style-type: none"> <li>* Has unknown antibody status, but was born to a mother known to be infected with HIV</li> </ul>
270.0	Disturbances of amino-acid transport Cystinosis Cystinuria Hartnup disease	0-20	Clinical history and physical exam; laboratory studies supporting diagnosis. Subspecialist consultation note may be required.
270.1	Phenylketonuria - PKU	0-20	Clinical history and physical exam; laboratory studies supporting diagnosis. Subspecialist consultation note may be required. Lab test: high plasma phenylalanine and normal/low tyrosine
270.2	Other disturbances of aromatic-acid metabolism	0-20	Clinical history and physical exam; laboratory studies supporting diagnosis. Subspecialist consultation note may be required.
270.3	Disturbances of branched-chain amino-acid metabolism	0-20	
270.4	Disturbances of sulphur-bearing amino-acid metabolism	0-20	
270.5	Disturbances of histidine metabolism Carnosinemia Histidinemia Hyperhistidinemia Imidazole aminoaciduria	0-20	Clinical history and physical exam; laboratory studies supporting diagnosis. Subspecialist consultation note may be required.
270.6	Disorders of urea cycle metabolism	0-20	Clinical history and physical exam; laboratory studies supporting diagnosis. Subspecialist consultation note may be required.

## Rare and Expensive Disease List as of July 1, 1998

ICD-9 Code	Disease	Age Group	Guidelines
270.7	Other disturbances of straight-chain amino-acid Glucoglycinuria Glycinemia (with methylmalonic acidemia) Hyperglycinemia Hyperlysinemia Pipicolinic acidemia Saccharopinuria Other disturbances of metabolism of glycine, threonine, serine, glutamine, and lysine	0-20	Clinical history and physical exam; laboratory studies supporting diagnosis. Subspecialist consultation note may be required.
270.8	Other specified disorders of amino-acid metabolism Alaninemia Ethanolaminuria Glycoprolinuria Hydroxyprolinemia Hyperprolinemia Iminoacidopathy Prolinemia Prolinuria Sarcosinemia	0-20	Clinical history and physical exam; laboratory studies supporting diagnosis. Subspecialist consultation note may be required.
271.0	Glycogenosis	0-20	Clinical history and physical exam; laboratory studies supporting diagnosis. Subspecialist consultation note may be required.
271.1	Galactosemia	0-20	Clinical history and physical exam; laboratory studies supporting diagnosis. Subspecialist consultation note may be required.
271.2	Hereditary fructose intolerance	0-20	Clinical history and physical exam; laboratory studies supporting diagnosis. Subspecialist consultation note may be required.
272.7	Lipidoses	0-20	Clinical history and physical exam; laboratory studies supporting diagnosis. Subspecialist consultation note may be required.
277.0	Cystic fibrosis	0-64	Clinical history and physical exam; laboratory studies supporting diagnosis. Subspecialist consultation note may be required.
277.00	Cystic fibrosis w/o ileus	0-64	
277.01	Cystic fibrosis with ileus	0-64	
277.2	Other disorders of purine and pyrimidine metabolism	0-64	Clinical history and physical exam; laboratory studies supporting diagnosis. Subspecialist consultation note may be required. Demonstration of deficient enzyme such as: alpha-L-Iduronidase, Iduronosulfate sulfatase, Heparan sulfate sulfatase, N-Acetyl-alpha-D-glucosaminidase, Arylsulfatase B, Beta-Glucuronidase, Beta-Galactosidase, N-Aacetylhexosaminidase-6-SO4 sulfatase.
277.5	Mucopolysaccharidosis	0-64	
277.8	Other specified disorders of metabolism	0-64	Clinical history and physical exam; laboratory or imaging studies supporting diagnosis. Subspecialist consultation note may be required.

## Rare and Expensive Disease List as of July 1, 1998

ICD-9 Code	Disease	Age Group	Guidelines
284.0	Constitutional aplastic anemia	0-20	Clinical history and physical exam; laboratory studies supporting diagnosis. Subspecialist consultation note may be required.
286.0	Congenital factor VIII disorder	0-64	Clinical history and physical exam; laboratory studies supporting diagnosis. Subspecialist consultation note may be required.
286.1	Congenital factor IX disorder	0-64	
286.2	Congenital factor XI deficiency	0-64	
286.3	Congenital deficiency of other clotting factors	0-64	
286.4	von Willebrand's disease	0-64	
330	Cerebral degenerations in childhood	0-20	Clinical history and physical exam; laboratory or imaging studies supporting diagnosis. Subspecialist consultation note may be required.
330.0	Leukodystrophy	0-20	
330.1	Cerebral lipidoses	0-20	Clinical history and physical exam; laboratory or imaging studies supporting diagnosis. Subspecialist consultation note may be required.
330.2	Cerebral degenerations in generalized lipidoses	0-20	
330.3	Cerebral degeneration of childhood in other diseases classified	0-20	Clinical history and physical exam; laboratory or imaging studies supporting diagnosis. Subspecialist consultation note may be required.
330.8	Other specified cerebral degeneration in childhood	0-20	
330.9	Unspecified cerebral degeneration in childhood	0-20	
331.3	Communicating hydrocephalus	0-20	Clinical history and physical exam; imaging studies supporting diagnosis. Subspecialist consultation note may be required.
331.4	Obstructive hydrocephalus	0-20	
333.2	Myoclonus	0-5	Clinical history and physical exam. Subspecialist consultation note may be required.
333.6	Idiopathic torsion dystonia	0-64	Clinical history and physical exam; laboratory or imaging studies supporting diagnosis. Subspecialist consultation note may be required.
333.7	Symptomatic torsion dystonia	0-64	Clinical history and physical exam; laboratory or imaging studies supporting diagnosis. Subspecialist consultation note may be required.
333.90	Unspecified extrapyramidal disease and abnormal movement disorder	0-20	Clinical history and physical exam; laboratory or imaging studies supporting diagnosis. Subspecialist consultation note may be required.
334	Spinocerebellar disease	0-20	Clinical history and physical exam. Neurology consultation note.
334.0	Friedreich's ataxia	0-20	
334.1	Hereditary spastic paraplegia	0-20	
334.2	Primary cerebellar degeneration	0-20	
334.3	Cerebellar ataxia NOS	0-20	

## Rare and Expensive Disease List as of July 1, 1998

ICD-9 Code	Disease	Age Group	Guidelines
334.4	Cerebellar ataxia in other diseases	0-20	
334.8	Other spinocerebellar diseases NEC	0-20	
334.9	Spinocerebellar disease NOS	0-20	
335	Anterior horn cell disease	0-20	Clinical history and physical exam. Neurology consultation note.
335.0	Werdnig-Hoffmann disease	0-20	
335.1	Spinal muscular atrophy	0-20	
335.10	Spinal muscular atrophy NOS	0-20	
335.11	Kugelberg-Welander disease	0-20	
335.19	Spinal muscular atrophy NEC	0-20	
335.2	Motor neuron disease	0-20	
335.20	Amyotrophic lateral sclerosis	0-20	
335.21	Progressive muscular atrophy	0-20	
335.22	Progressive bulbar palsy	0-20	
335.23	Pseudobulbar palsy	0-20	
335.24	Primary lateral sclerosis	0-20	
335.29	Motor neuron disease NEC	0-20	
335.8	Anterior horn disease NEC	0-20	
335.9	Anterior horn disease NOS	0-20	
341.1	Schilder's disease	0-64	Clinical history and physical examination; supporting imaging studies and neurologic consultation note may be required.
343.0	Diplegic infantile cerebral palsy	0-20	Clinical history and physical examination; supporting imaging studies and neurologic consultation note may be required.
343.2	Quadriplegic infantile cerebral palsy	0-64	Clinical history and physical examination; supporting imaging studies and neurologic consultation note may be required.
344.0	Quadriplegia	0-64	Clinical history and physical examination; supporting imaging studies and neurologic consultation note may be required.
359.0	Congenital hereditary muscular dystrophy	0-64	Clinical history and physical examination; supporting imaging studies and neurologic consultation note may be required.
359.1	Hereditary progressive muscular dystrophy	0-64	Clinical history and physical examination; supporting imaging studies and neurologic consultation note may be required.
359.2	Congenital myotonic dystrophy (Steinert's only)	0-64	Clinical history and physical examination; supporting imaging studies and neurologic consultation note may be required.
437.5	Moyamoya disease	0-64	Clinical history and physical examination; supporting imaging studies and neurologic consultation note may be required.



## Rare and Expensive Disease List as of July 1, 1998

ICD-9 Code	Disease	Age Group	Guidelines
579.3	Short gut syndrome	0-20	Clinical history and imaging studies supporting diagnosis. Gastrointestinal subspecialist consultation note may be required.
582	Chronic glomerulonephritis	0-20	Clinical history, laboratory evidence of renal disease. Nephrology subspecialist consultation note may be required.
582.0	Chronic glomerulonephritis with lesion of proliferative glomerulonephritis	0-20	Clinical history, laboratory evidence of renal disease. Nephrology subspecialist consultation note may be required.
582.1	Chronic glomerulonephritis with lesion of membranous glomerulonephritis	0-20	Clinical history, laboratory evidence of renal disease. Nephrology subspecialist consultation note may be required.
582.2	Chronic glomerulonephritis with lesion of membranoproliferative glomerulonephritis	0-20	Clinical history, laboratory evidence of renal disease. Nephrology subspecialist consultation note may be required.
582.4	Chronic glomerulonephritis with lesion of rapidly progressive glomerulonephritis	0-20	Clinical history, laboratory evidence of renal disease. Nephrology subspecialist consultation note may be required.
582.8	Chronic glomerulonephritis with other specified pathological lesion in kidney	0-20	Clinical history, laboratory evidence of renal disease. Nephrology subspecialist consultation note may be required.
582.81	Chronic glomerulonephritis in diseases classified elsewhere	0-20	Clinical history, laboratory evidence of renal disease. Nephrology subspecialist consultation note may be required.
582.89	Other Chronic glomerulonephritis with lesion of exudative nephritis interstitial (diffuse) (focal) nephritis	0-20	Clinical history, laboratory evidence of renal disease. Nephrology subspecialist consultation note may be required.
582.9	With unspecified pathological lesion in kidney Glomerulonephritis: NOS specified as chronic hemorrhagic specified as chronic Nephritis specified as chronic Nephropathy specified as chronic	0-20	Clinical history, laboratory evidence of renal disease. Nephrology subspecialist consultation note may be required.
585	Chronic renal failure A) diagnosed by a pediatric nephrologist	0-20	Clinical history, laboratory evidence of renal disease. Pediatric nephrology subspecialist consultation note required.
585. V45.1	B) with dialysis and documented rejection from Medicare	21-64	Clinical history, laboratory evidence of renal disease. Nephrology subspecialist consultation note may be required.
741	Spina bifida	0-64	Clinical history and physical exam. Imaging studies supporting diagnosis. Subspecialist consultation note may be required.
741.0	Spina bifida with hydrocephalus	0-64	
741.00	Spina bifida with hydrocephalus NOS	0-64	
741.01	Spina bifida with hydrocephalus cervical region	0-64	
741.02	Spina bifida with hydrocephalus dorsal region	0-64	

## Rare and Expensive Disease List as of July 1, 1998

ICD-9 Code	Disease	Age Group	Guidelines
741.03	Spina bifida with hydrocephalus lumbar region	0-64	
741.9	Spina bifida without hydrocephalus	0-64	
741.90	Spina bifida unspecified region	0-64	
741.91	Spina bifida cervical region	0-64	
741.92	Spina bifida dorsal region	0-64	
741.93	Spina bifida lumbar region	0-64	
742.0	Encephalocele Encephalocystocele Encephalomyelocele Hydroencephalocele Hydromeningocele, cranial Meningocele, cerebral Menigoencephalocele	0-20	Clinical history and physical examination, radiographic or other neuroimaging studies. Neurology or neurosurgery consultation note may be required.
742.1	Microcephalus Hydromicrocephaly Micrencephaly	0-20	Clinical history and physical examination, radiographic or other neuroimaging studies. Neurology or neurosurgery consultation note may be required.
742.3	Congenital hydrocephalus	0-20	Clinical history and physical examination, radiographic or other neuroimaging studies. Neurology or neurosurgery consultation note may be required.
742.4	Other specified anomalies of brain	0-20	Clinical history and physical examination, radiographic or other neuroimaging studies. Neurology or neurosurgery consultation note may be required.
742.5	Other specified anomalies of spinal cord	0-64	Clinical history and physical examination, radiographic or other neuroimaging studies. Neurology or neurosurgery consultation note may be required.
742.59	Other specified anomalies of spinal cord Amyelia Congenital anomaly of spinal meninges Myelodysplasia Hypoplasia of spinal cord	0-64	Clinical history and physical examination, radiographic or other neuroimaging studies. Neurology or neurosurgery consultation note may be required.
748.1	Nose anomaly - cleft or absent nose ONLY	0-5	Clinical history and physical examination. Radiographic or imaging studies and specialist consultation note (ENT, plastic surgery) may be required.
748.2	Web of larynx	0-20	Clinical history and physical exam; laboratory or imaging studies supporting diagnosis. Subspecialist consultation note may be required.
748.3	Laryngotracheal anomaly NEC- Atresia or agenesis of larynx, bronchus, trachea, only	0-20	
748.4	Congenital cystic lung	0-20	Clinical history and physical exam; imaging studies supporting diagnosis. Subspecialist consultation note may be required
748.5	Agenesis, hypoplasia and dysplasia of lung	0-20	

## Rare and Expensive Disease List as of July 1, 1998

ICD-9 Code	Disease	Age Group	Guidelines
749 except 749.1x	Cleft palate and cleft lip	0-20	Clinical history and physical examination. Supporting consultation note from ENT/plastic surgery may be required.
749.0	Cleft palate	0-20	Clinical history and physical examination. Supporting consultation note from ENT/plastic surgery may be required.
749.00	Cleft palate NOS	0-20	
749.01	Unilateral cleft palate complete	0-20	
749.02	Unilateral cleft palate incomplete	0-20	
749.03	Bilateral cleft palate complete	0-20	
749.04	Bilateral cleft palate incomplete	0-20	
749.2	Cleft palate with cleft lip	0-20	
749.20	Cleft palate and cleft lip NOS	0-20	
749.21	Unilateral cleft palate with cleft lip complete	0-20	
749.22	Unilateral cleft palate with cleft lip incomplete	0-20	
749.23	Bilateral cleft palate with cleft lip complete	0-20	
24	Bilateral cleft palate with cleft lip incomplete	0-20	
749.25	Cleft palate with cleft lip NEC	0-20	
750.3	Congenital tracheoesophageal fistula, esophageal atresia and stenosis	0-3	Clinical history, physical examination; imaging studies supporting diagnosis. Subspecialist consultation note may be required.
751.2	Atresia large intestine	0-5	Clinical history and physical exam; laboratory or imaging studies supporting diagnosis. Subspecialist consultation note may be required.
751.3	Hirschsprung's disease	0-15	
751.61	Biliary atresia	0-20	
751.62	Congenital cystic liver disease	0-20	
751.7	Pancreas anomalies	0-5	
751.8	Other specified anomalies of digestive system NOS	0-10	
753.0	Renal agenesis and dysgenesis, <b>bilateral only</b> Atrophy of kidney: congenital infantile Congenital absence of kidney(s) Hypoplasia of kidney(s)	0-20	Clinical history, physical examination, radiographic or other imaging studies. Subspecialist consultation note may be required.
753.1	Cystic kidney disease, <b>bilateral only</b>	0-20	Clinical history, physical examination, radiographic or other imaging studies. Subspecialist consultation note may be required.

## Rare and Expensive Disease List as of July 1, 1998

ICD-9 Code	Disease	Age Group	Guidelines
753.12	Polycystic kidney, unspecified type, <b>bilateral only</b>	0-20	Clinical history, physical examination, radiographic or other imaging studies. Subspecialist consultation note may be required.
753.13	Polycystic kidney, autosomal dominant, <b>bilateral only</b>	0-20	Clinical history, physical examination, radiographic or other imaging studies. Subspecialist consultation note may be required.
753.14	Polycystic kidney, autosomal recessive, <b>bilateral only</b>	0-20	Clinical history, physical examination, radiographic or other imaging studies. Subspecialist consultation note may be required.
753.15	Renal dysplasia, <b>bilateral only</b>	0-20	Clinical history, physical examination, radiographic or other imaging studies. Subspecialist consultation note may be required.
753.16	Medullary cystic kidney, <b>bilateral only</b>	0-20	Clinical history, physical examination, radiographic or other imaging studies. Subspecialist consultation note may be required.
753.17	Medullary sponge kidney, <b>bilateral only</b>	0-20	Clinical history, physical examination, radiographic or other imaging studies. Subspecialist consultation note may be required.
753.5	Exstrophy of urinary bladder	0-20	Clinical history, physical examination, radiographic and/or other imaging studies. Subspecialist consultation note may be required.
756.0	Musculoskeletal--skull and face bones Absence of skull bones Acrocephaly Congenital deformity of forehead Craniosynostosis Crouzon's disease Hypertelorism Imperfect fusion of skull Oxycephaly Platybasia Premature closure of cranial sutures Tower skull Trigonocephaly	0-20	Clinical history, physical examination; imaging studies supporting diagnosis. Subspecialist consultation note may be required.
756.4	Chondrodystrophy	0-1	Clinical history and physical exam; imaging studies supporting diagnosis. Subspecialist consultation note may be required.
756.50	Osteodystrophy NOS	0-1	Clinical history and physical exam; imaging studies supporting diagnosis. Subspecialist consultation note may be required.
756.51	Osteogenesis imperfecta	0-20	Clinical history, physical examination, radiologic studies. Specialist consultation report (genetics, orthopedics) may be required.
756.52	Osteopetrosis	0-1	Clinical history and physical exam; imaging studies supporting diagnosis. Subspecialist consultation note may be required.
756.53	Osteopoikilosis	0-1	

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ICD-9 Code	Disease	Age Group	Guidelines
756.54	Polyostotic fibrous dysplasia of bone	0-1	
756.55	Chondroectodermal dysplasia	0-1	
756.56	Multiple epiphyseal dysplasia	0-1	
756.59	Osteodystrophy NEC	0-1	
756.6	Anomalies of diaphragm	0-1	Clinical history and physical exam; imaging studies supporting diagnosis. Subspecialist consultation note may be required.
756.7	Abdominal wall anomalies	0-1	Clinical history and physical exam.
759.7	Multiple congenital anomalies NOS	0-10	Clinical history and physical exam; laboratory or imaging studies supporting diagnosis. Subspecialist consultation note may be required.
V46.1	Dependence on respirator	1-64	Clinical history and physical exam. Specialist consultation note required.
V46.9	Machine dependence NOS	1-64	

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May 17, 1999